

almost a week no more bleeding occurred. It then began again as profusely as before, so the kidney was removed and the hæmaturia ceased. Macroscopically the kidney revealed no evidence to account for the bleeding, but of course showed an infarct corresponding to the divided aberrant artery.

Microscopically Dr. Davis found degeneration of parenchyma and glomeruli, and the tubules denuded of their epithelium, with catarrhal inflammation. Even this seems inadequate to account for such severe hæmorrhage.

SUMMARY.

In the absence of a calculus or infective condition, a neoplasm seemed the most probable cause of severe hæmaturia in a patient of his age. The absence of a filling defect in the retrograde pyelograms would seem to exclude a tumour encroaching on the pelvis, but would not dispose of the possibility of a small papilloma of the pelvis. The first operative finding appeared to provide a possible cause, and was provisionally accepted in view of the fact that in cases of so-called "essential hæmaturia" the remaining kidney may start to bleed when the primary bleeding one has been removed. This condition is, however, nearly always a hæmorrhagic nephritis, and it is probable that some evidence of abnormality would be revealed in both by the uroselectan test. The possibility of an association with his asthma remains.

This patient was seen in consultation with Dr. C. W. Kidd, to whom I am indebted for permission to publish the case.

REPORT ON CASE OF HÆMOCHROMATOSIS OR BRONZED DIABETES

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*With Post-Mortem Report by J. A. Fisher, M.D., and Special Report
on the Iron Content of the Tissues by S. Andrews, M.B., B.Sc.*

INTRODUCTORY.

THIS disease is characterized by three outstanding features :—

1. Accumulation of iron in the body, with pigmentation of the skin and internal organs.
2. Cirrhosis of the liver.
3. Fibrosis of the pancreas with or without glycosuria.

It was formerly supposed that hæmochromatosis was due to a primary blood disease involving the breaking down of hæmoglobin and the deposition of iron, which caused irritation and a secondary sclerosis of the tissues in which it was deposited. It is now generally believed that the increased iron content of the tissues is the result of some abnormality in the metabolism of iron, whereby the iron is retained in the tissues and not excreted.

The case which we are now recording is very similar to one which I published in the Transactions of the Ulster Medical Society in 1927. In that paper I described the pigmentation as a dull leaden tinting of the skin, most marked on the face and on the back of the hands.

Hanot and Chauffard have described the pigmentation as a uniform leaden tint, dull earthy, but with a grey rather than a brown hue; something, in a word, absolutely characteristic. Sheldon called it a bluish-bronze colour. Barth states: "It is not a frankly bronzed discoloration, but rather a shade of blackish-grey with metallic tints reminiscent of cast iron."

These descriptions show that bronze is not the usual colour of the pigmentation, though possibly in some cases there may have been jaundice super-added. Bronze is generally accepted as being a reddish-brown colour.

I have since seen five other cases, in all of which the pigmentation was of a leaden tint and very characteristic. The term 'bronze' would not be a correct description of the colour of any of these cases.

NOTES OF PRESENT CASE.

Male, aged 38, police constable. Married for three years; no children. Admitted to hospital on the 4th April, 1935.

Family history.—There was no record of the occurrence of diabetes or of abnormal pigmentation.

Personal history.—He enjoyed good health up till 1928, when he developed diabetes, and had been taking insulin daily since then. He was not sure when the pigmentation commenced, but had noticed it for about two years.

History of present affection.—About two weeks prior to admission his feet and legs became swollen, his abdomen distended, and he was troubled with shortness of breath on exertion.

Condition on admission.—Patient was tall and of spare build. Nutrition was poor. Skin was of a peculiar slate colour, most marked on the face and hands (this description of the pigmentation was noted by my house physician). Growth of hair on the scalp was normal. Face smooth: he only required to shave once weekly. Pubic hair was thin and very scanty. Testicles were small. Oedema of feet and ankles was present.

Heart and lungs were normal. Pulse was 84. Arteries were not sclerosed. Blood-pressure was 98/70 mm. Hg.

Abdomen.—The liver was enlarged to the level of the umbilicus; it was firm and not tender. Free fluid was present in the peritoneal cavity. The spleen was palpable.

Nervous system.—Knee jerks and ankle jerks were absent, and nothing else abnormal was noted.

The urine contained sugar and acetone, but no albumen. Blood-sugar on admission was 0.37 per cent. Wassermann reaction was negative.

His general condition improved under treatment, the ascites and oedema almost disappeared, and his weight fell from 11st. 2lb. to 9st. 8lb. The blood-sugar was difficult to control, as he had frequent hypo-glycæmic reactions. He was discharged on the 16th June, taking forty units of insulin daily.

He was readmitted on the 26th August with a recurrence of his symptoms, and improved for a time, but gradually became weaker, and died on the 1st November, 1935. There was no marked somnolence or mental dullness present until a few days before death.

POST-MORTEM EXAMINATION.

The body was that of a poorly nourished male subject, and exhibited a generalized greyish-brown pigmentation. The sclerotics were bluish-white in colour; there was no enlargement of the superficial lymph-glands; there was slight œdema of the feet; there was normal growth of hair on the scalp, but pubic hair was exceedingly scanty. The testes were very small. Healed pigmented scar of old varicose ulcer was seen over the middle third of left tibia. On the anterior aspect of the thigh on the right side was a healed surgical scar two and a half inches long, which was white in colour, in contrast to the surrounding pigmented skin.

Thorax.—On reflecting the skin and superficial fascia, the intercostal muscles were seen to be brownish-red in colour—the brown colour of these muscles being very striking when compared with those of another body on which a post-mortem examination had just been performed.

There was approximately half a pint of clear yellow fluid in each pleural sac. The lungs were œdematous and were the seat of a broncho-pneumonic consolidation affecting the lower lobes.

The pericardial sac contained an excess of clear yellow fluid.

The heart weighed 1lb. 2oz. The muscle was flabby in consistence and rusty brown in colour. The left ventricle was enlarged as the result of dilatation of the cavity. Apart from a few small patches of fatty degeneration on the ventricular surface of the aortic valves and anterior cusp of mitral valve, the heart presented no abnormalities.

Abdomen.—The peritoneal sac contained a large quantity of clear yellow fluid. There were a few calcified mesenteric glands in the mesentery of the lower part of the ileum. There were a number of enlarged brown, fleshy lymph-glands in the retro-peritoneal tissue, most marked in the upper part of the abdomen.

The liver was enlarged and weighed 7lb. It was brownish-red in colour, and was the seat of a multilobular cirrhosis. Its surface was stippled with small white nodules, representing foci of hyperplasia.

The stomach was dilated. There was a brown pigmentation of its wall, seen best by holding the stomach up to the light. The duodenum was healthy.

The pancreas weighed 4oz. It was soft in consistence and deep brown in colour.

The kidneys weighed 10oz. each. On section, apart from this enlargement and some congestion, they showed no gross abnormality.

Adrenals were healthy in appearance.

The spleen weighed 8oz. No notable changes were seen in pulp. Fibrous trabeculae were thickened.

The prostate was diminished in size, and the urinary bladder healthy.

Testes were small, soft in consistence, and rather brown on section.

MICROSCOPICAL EXAMINATION.

The liver was the seat of a multilobular cirrhosis. There was a very large amount of iron pigment contained within the liver-cells and also dispersed widely through the interstitial tissue. Smaller granules of the same pigment were observed in the epithelium of the bile-ducts. It was noteworthy that the regenerating liver-cells in the nodules of hyperplasia were free from pigment.

The pancreas exhibited a well marked fibrosis. The glandular elements were degenerated to such an extent that the islets of Langerhans could not be identified. The degenerated parenchymatous cells contained much hæmosiderin.

The lymph-glands from the upper part of the abdomen contained a large amount of hæmosiderin, for the most part contained in phagocytes. The pigment occurred in localized collections, and in these areas an increase of fibrous tissue had occurred. A few foreign body giant cells were present.

The skin showed fine deposits of hæmosiderin in relation to the sweat glands.

The muscle fibres of the heart were loaded with granules of hæmosiderin, which were situated mainly around the poles of the nuclei.

Testis.—The seminiferous tubules and the interstitial cells of Leydig had undergone an extreme degree of atrophy associated with a remarkable increase of loose interstitial tissue. No iron could be demonstrated in the tissues of this gland.

IRON CONTENT OF THE TISSUES.

The amount of iron in some of the organs is given in the table below. The results are expressed as percentage of the dry weight. Analyses of a former case (November, 1927) are also given, since they have not previously been recorded. For comparison, the normal values for iron are given, and also the average values for all the recorded cases of the disease (Sheldon).

<i>Organ</i>	PERCENTAGE OF DRY WEIGHT.				
	<i>Present Case</i>	<i>Case November, 1927</i>	<i>Normal Values</i>	<i>Average of Recorded Cases</i>	
Liver - -	3.330	3.740	0.075	3.650	
Heart - -	0.530	0.329	0.039	0.517	
Spleen - -	0.363	1.390	0.140	0.631	
Kidney - -	0.145	0.140	0.039	0.195	
Pancreas - -	—	1.200	0.018	1.890	

The large accumulation of iron which takes place in the body in this disease is illustrated by the above figures. The total amount of iron in the whole human body is normally about 5 to 8 gms., and the average daily intake in the food is 10 to 20 mgms. Calculating from the analysis of the liver in the present case (weight 7lb.), we find that this organ alone contains actually 26.4 gms. of iron. Making a very rough approximate calculation, we might say that the whole body contained between 50 and 60 gms. of the metal, which would mean, with a daily intake and complete retention of the 10 mgms. in the food, that there was a gradual accumulation of the iron over about fifteen years.